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In this edition

PEDIATRIC NEWS

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CHIEF'S CORNER

Pediatric Imaging Services

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The Pediatric Imaging Service at Wilford Hall Medical Center currently consists of 3 fellowship trained pediatric radiologists: Dr. Randy Richardson, Dr. Chris Keesling and Dr. Cindy Taylor. State-of -the-art diagnostic imaging examinations, in all modalities, are available for our pediatric population. Recently, we have also added the capability for Trans-cranial Doppler examinations and ultrasound evaluation of intussusceptions. To ensure our smallest patients receive the correct preparation/examination and to minimize radiation dose, consultation with the pediatric radiologist is encouraged. Understanding that many cases present after normal duty hours, a pediatric radiologist is always available (24/7) to provide expertise in the appropriate imaging workup and diagnosis.

Scheduling of pediatric patients is relatively simple. Order entry into CHCS is essential in all cases. In most cases, our scheduling office will call the patient's family with the appointment time. In general, appointments can be made within a few days although some, usually MRIs (see below), have a waiting time of several weeks. More urgent cases and hospitalized patients can be added to an existing schedule after consultation with and approval by the pediatric radiologist.

Many examinations, mostly MRI scans, require sedation. The pediatric anesthesiologist performs the majority of sedations and there are limited appointments for these cases. Unfortunately, many patients are turned away from these appointments due to improper preparation, subsequently, these scheduled slots go unused. Review of the sedation preparation instructions with the patients and their parents may be beneficial in maximizing utilization of these valuable appointments. Coordination with the credentialed provider performing the sedation and the imaging center performing the examination is also essential. Only providers certified to perform conscious sedation IAW 44-21 will be allowed to administer sedation in the radiology department.

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When Should a Head CT Scan Be Done Before a Lumbar Puncture for Bacterial Meningitis?

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This concern regularly arises. Adult neurologists are taught to routinely do a head CT scan before an LP, unless there are clearcut signs of normal intracranial pressure, such as the presence of venous pulsations, no papilledema, and no focal neurologic signs.

Robert A. Fishman writes in Merritt's Neurology that a serious complication of a lumbar puncture is the possibility of aggravating a preexisting, often unrecognized brain herniation syndrome (e.g., uncal, cerebellar, or cingulate herniation) associated with intracranial hypertension. This hazard is the basis for considering papilledema to be a relative contraindication to a lumbar puncture. In the presence of papilledema, if a head CT reveals no evidence of a mass lesion or edema, then an LP can be safely done.

Fishman also writes in his own book Cerebrospinal Fluid in Diseases of the Nervous System, that a head CT scan and MR imaging have simplified the management of patients with papilledema and suspected intracranial hypertension and altered states of consciousness. These techniques establish the absence or presence of distortion or displacement of the ventricular system, sulcal effacement, and tentorial, uncal, or cerebellar herniation, which facilitates the physician's decision regarding the appropriateness of a spinal puncture. Such changes, particularly when severe, are generally considered as contraindications to a lumbar puncture.

In a child suspected to have meningitis, it is frequently difficult to detect papilledema. Early signs of papilledema usually shows after 48 hours of persistent increased intracranial pressure, so it may not even be present. So how can a pediatrician determine whether or not it is safe for a child to have a lumbar puncture performed in a suspected case of meningitis? It is well known, as well as, well documented that bacterial meningitis is associated with increased intracranial pressure. Why else is the open fontanelle bulging?

It is written in Pediatric Emergency Medicine, Concepts and Clinical Practice, that if there is any question of increased intracranial pressure on the basis of history and physical examination, a CT scan should be obtained before the lumbar puncture is performed. The APLS Pediatric Emergency Medicine Course writes that in selected cases, a CT scan may be warranted before a lumbar puncture. A child with focal neurologic findings and findings suggestive of increased intracranial pressure should have a CT scan before a lumbar puncture. Finally, Robert M. Reece, M.D., writes in the 1992 Manual of Emergency Pediatrics that the patient who presents with sepsis or meningitis requires a rapid workup and initiation of therapy at the earliest opportunity for the most optimal outcome. A CT scan of the head should be performed prior to the LP in patients with suspected increased intracranial pressure or intracranial mass lesion.

So, it seems clear in both adult neurology references and pediatric emergency medicine references that a CT scan needs to be done before a lumbar puncture for meningitis evaluation, if there are signs and symptoms of increased intracranial pressure, or focal neurologic signs. The guidelines seem simple and clearcut. But, nowhere is it written that an open fontanelle takes away the risk of cerebral herniation. And what is to be done in light of a child who is sluggish because of a fever, who has a bulging fontanelle? Both of these conditions are indications for an LP, looking for meningitis in a child under 12 months of age. At the same time, both of these are clinical manifestations of increased intracranial pressure. Strict adherence to the above mentioned guidelines strongly suggest a head CT scan be done before the lumbar puncture is to be performed.

David H. Mellor, M.D., concludes that "there is no place for routine computed tomography before or after lumbar puncture in the child with clinically uncomplicated acute bacterial meningitis." So, is a febrile, sleepy 6 month old child with a bulging fontanelle a case of a "clinically uncomplicated acute bacterial meningitis"? Many of us older pediatricians have done lumbar punctures many times in such children, finding many cases of acute bacterial meningitis, without doing a CT scan. Of course, the lumbar puncture would be withheld if the child was too septic, there was evidence of coagulopathy, or there was an overlying skin infection.

Abnormalities of the head CT scan, suggesting increased intracranial pressure, involves the brain parenchyma, the ventricles, the cisterns, the sutures, and the foramen magnum. Cerebral edema in the brain parenchyma would show up as increased density of the cerebrum, basal ganglia, and cortex, and loss of the grey-white differen-

tiation. Pediatric radiologists can also look at the cerebrum to cerebellum ratio to add to the suspicion of cerebral edema. Increased intracranial pressure can show up as enlargement of any of the 4 ventricles. The decreased size of the suprasellar, prepontine, cerebellopontine, midbrain cisterns and of the cisterna magna would suggest cerebral edema. Splitting of the sutures on the bone films, or a full foramen magnum, would suggest increased intracranial pressure.

There was a recent case reviewed for risk management, involving the above circumstances. A 6 month old child presented to a military emergency room with a history of fever and some sleepiness, a bulging fontanelle, perhaps some irritability and a decreased appetite. A lumbar puncture was ordered, but a head CT scan was done first. It showed signs of some ventriculomegaly. A neurologist was consulted, and the go ahead was given for the lumbar puncture to proceed. The results showed purulent meningitis, the child was immediately treated with antibiotics, but within several hours, clinically deteriorated due to cerebral herniation. In retrospect, one can say that there were several signs of increased intracranial pressure, with the bulging fontanelle, with the sleepiness, with the ventriculomegaly. Would you have done the CT scan in the first place? How could you disprove in court that the lumbar puncture greatly contributed to herniation of the brain?

There are studies to show that cerebral herniation occurs in 4-6% of cases of bacterial meningitis in industrialized nations, and as much as 15% of cases of bacterial meningitis in Third World countries. Rennick, et al, reports that in a study of 445 children over 30 days of age, from 1984 to 1989, cerebral herniation occurred in 19 of the children (4.3%). And 14 of the 19 children (74%) with herniation died. Interestingly, a head CT scan was performed at about the time of herniation on 14 occasions, and 5 of the tomograms yielded normal results (36%). The five tomograms with normal results were from four children. Two of the children had herniation confirmed at postmortem examination, and the other two survived. So even with a normal head CT scan, cerebral herniation can still occur.

This same study recorded that eight (38%) of the 21 episodes of cerebral herniation occurred within 3 hours of the lumbar puncture, and 12 (57%) occurred within 12 hours of the lumbar puncture. Six (29%) of the 21 episodes of herniation occurred before lumbar puncture was performed, or in a child who did not undergo puncture. The clear temporal relation between lumbar puncture and herniation strong suggests that the procedure causes herniation in some patients. Horwitz et al found that eight out of 18 cases of herniation occurred within 30 minutes of admission, suggesting that lumbar puncture may have been the cause. These 18 cases of herniation occurred in 6% of a retrospective study of 302 infants and children with pyogenic meningitis. Four of the 15 patients who survived were left with severe residua (hemiparesis, cortical blindness, seizures, and mental retardation). Dodge and Swartz found that herniation occurred within two hours of lumbar puncture in three out of nine cases diagnosed at postmortem examination. Rosenberg et al, however, found no relation between lumbar puncture and herniation.

In one study from Nigeria, where head CT scans are not readily available, signs of cerebral herniation occurred in 10% of patients. Even when Mannitol was used successfully to resolve the signs and symptoms of cerebral herniation in 11 patients, 4 ended up having cerebral herniation after the lumbar puncture. More frighteningly, 8% of patients without any signs of cerebral herniation , ended up having herniation occur after the lumbar puncture. In this study, possible herniation was defined as the presence of at least two of four clinical criteria: unarousable coma, unilateral or bilateral dilated or unreactive pupils, abnormal pattern of respiration (hyperventilation, irregular respiration, apnea), and abnormal posturing (decorticate or decerebrate) or complete flaccidity.

The following conditions are contraindications to performing a lumbar puncture, in a child with suspected acute bacterial meningitis: papilledema, coma, hypertension, bradycardia, bradypnea or irregular respirations, fixed dilated, or unequal pupils, absent doll's eye movements, recent or prolonged convulsive seizures, tonic seizures, decerebrate or decorticate posturing, hemiparesis, septic shock. In such cases, parenteral antibiotics should not be delayed much longer than it takes to draw blood for a blood culture.

So, let us review the guidelines again that are in agreement from many sources.

A lumbar puncture should be withheld in the event of increased intracranial pressure or focal neurologic signs. Then, perform a head CT scan prior to a lumbar puncture looking for radiologic evidence of increased intracranial pressure. Absolute contraindications to a lumbar puncture include coagulopathy, respiratory and circulatory compromise, and an overlying skin infection.

Appropriate questions that arise from clinical situations include:

Does a febrile child with a bulging fontanelle require a head CT scan before the lumbar puncture, since the bulging fontanelle suggests increased intracranial pressure? And does the sleepiness of such a child come from the fever, or from meningitis, or from the increased intracranial pressure? *Clinical judgement should be used, rather than absolute guidelines.* .

Does a normal head CT scan convincingly suggest that there is no increased intracranial pressure? *The study by Rennick quoted above shows that of 14 cases with signs of increased intracranial pressure when a CT scan was done, 5 of those cases had a normal head CT scan, and still ended up cerebral herniation.*

If increased intracranial pressure is the rule rather than the exception in bacterial meningitis, is it too risky to perform an LP in convincing cases of bacterial meningitis? *Multiple studies suggest that cerebral herniation occurs in 4-6% of cases of pyogenic meningitis in industrialized nations, whether or not a lumbar puncture was done.*

The only reasonable conclusions would include the following:

1. There should be no place for routine computed tomography before or after lumbar puncture in the child with clinically uncomplicated acute bacterial meningitis.
2. If increased intracranial pressure is undoubtedly present with moderate to severe signs and symptoms, then the lumbar puncture should be deferred, the patient be given parenteral antibiotics, after blood cultures are obtained, and the patient be sent to the ICU.
3. Clinical judgement needs to be used for other cases. Each physician must judge for himself or herself what constitutes an uncomplicated acute bacterial meningitis. If ever in doubt, always err on the side of caution and obtain the head CT scan before the lumbar puncture.
4. Even if the head CT scan is normal, if the lumbar puncture is strongly suggestive of a bacterial meningitis, be on alert for the possibility of cerebral herniation, which apparently occur in 4-6% of cases of bacterial meningitis.

Another way of looking at the problem, would be as follows:

1. If the child is too sick, defer the LP, treat, admit to the ICU.
2. If the child is too sick to be discharged from the ER, even with a normal LP, but not sick enough to be in an ICU, then do the head CT scan before the lumbar puncture.
3. If the child looks well enough to go home at presentation, and if the lumbar puncture would prove to be negative, then the head CT scan need not be done before the lumbar puncture, unless there are focal neurologic signs.
4. Any child with documented bacterial meningitis, needs to be carefully watched for impending cerebral herniation in the first 48 hours of treatment.

References:

Addy DP, (1987) when not to do a lumbar puncture. *Arch Dis Ch* 62:873-5.

Akpede GO, and Ambe JP, Cerebral herniation in pyogenic meningitis: prevalence and related dilemmas in emergency room populations in developing countries. *Dev Med & Ch Neuro* 2000, 42: 462-469.

Brown JK. (1976) Lumbar puncture and its hazards. *Dev Med & Ch Neurology* 18: 803-6\

Dodge PR, Swartz MN. Bacterial meningitis—a review of selected aspects. II. Special neurologic problems, postmeningitic complications and clinicopathological correlations. *N Engl J Med* 1965; 272: 954-60

Fishman, Robert A., Merritt's Neurology (10th ed.), ed. by Lewis P. Rowland, Lippincott Williams & Wilkins, 2000, p. 89.

Fishman, Robert A., Cerebrospinal Fluid in Diseases of the Nervous System, (2nd ed.), WB Saunders, 1992, p. 172.

Horwitz, SJ, Boxerbaum B, O'Bell J, Cerebral herniation in bacterial meningitis in childhood *Ann Neurol* 1980; 7:524-8

Mellor DH, The Place of Computed Tomography and Lumbar Puncture in Suspected Bacterial Meningitis, *Arch Dis Ch*, 67: 1417-9

Reece, Robert M., Manual of Emergency Pediatrics, WB Saunders, 1992, p. 74.

Rennick G, Shann F, de Campo J, Cerebral herniation during bacterial meningitis in children, *BMJ*, 1993, 306: 953-5.

Rosenberg DI, McCrory JH, Abhoudon MK, Murante AA, Downs CE. Neuro-intensive care for cerebral herniation in childhood meningitis. In: Hoff Jt, Betz AL, eds. *Intracranial pressure VII*. Berlin: Springer 1989: 770-2 (proceedings of the seventh interantional symposium on intracranial pressure

APLS The Pediatric Emergency Medicine Course, (3rd ed.), ed. by

Gary R. Strange, 1998, p. 178-9

Pediatric Emergency Medicine, (2nd ed.), ed. by Roger M. Barkin, Mosby, 1997, p. 999.

Central Auditory Processing Disorders

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How many times have you heard from a parent, "Little Tommy's teacher says he is hyperactive and needs to be on Ritalin." This has probably happened more times than you care to acknowledge. Central Auditory Processing Disorder (CAPD) and ADHD have many of the same characteristics and may coexist or be mutually exclusive of one another. CAPD should be taken into consideration before making the diagnosis of ADHD.

Children with CAPD can display similar behaviors as those children diagnosed with ADHD. Tillery and Keller, 1999 list the following as examples: requiring repetition of directions, acting as if a hearing loss is present, inability to understand complex/lengthy directions, poor reading comprehension, inconsistent academic performance, distractibility, language disorders, short term memory difficulties, inability to listen in background noise, noticeable improvement in performance in a structured environment, inattentiveness, and hyperactivity or hypoactivity.

Taking the time to rule out other factors contributing to a child's attention, activity, learning, and/or behavior problems may prevent a child from being labeled ADHD. This paper will focus on the risk factors, characteristics, testing available, and management techniques for those children diagnosed with CAPD.

The American Speech-Language-Hearing Association (ASHA) Task Force on Central Auditory Processing Consensus Development (1996) has defined auditory processing disorder as "a deficiency in one or more of the following phenomena: sound localization and lateralization, auditory discrimination, auditory pattern recognition, recognition of temporal aspects of audition, auditory performance decrease with competing acoustic signals, and auditory performance decrease with degraded signals." Basically CAPD is the inability to understand spoken language in a meaningful way.

There are important risk factors and characteristics of CAPD that would indicate a need for referral to an audiologist and speech pathologist. Information gained from these two disciplines will assist in determining whether a child has CAPD and what areas of strength and weakness are seen in the auditory processing system. This information can also be used to determine a management plan that best suits a child's needs.

Risk factors that need to be considered include a family history of CAPD, academic and attention problems. A history of chronic otitis media during early childhood, and the duration of each episode is another risk factor.

Characteristics of CAPD also include speech and language difficulties, problems with reading and spelling, phonological awareness, auditory memory, auditory sequencing, and auditory discrimination.

Speech pathologists utilize many tests to assess auditory processing skills. These tests include evaluating auditory perceptual skills which includes: selective attention, discrimination, recognition memory, memory for content, memory for sequence, sound mimicry, recognition, analysis, blending, sound-symbol association, reading of symbols, and spelling of sounds, sentence memory, and word memory. Other tests that evaluate auditory-based capabilities include: auditory discrimination tests, receptive and expressive language tests, and tests of written language abilities.

The audiologist's testing can be quite extensive depending on the CAPD characteristics present in a child. These tests will provide a comprehensive assessment of a child's central auditory processing skills and areas of strength and weakness. Testing should include dichotic speech tests, temporal ordering tasks, monaural low-redundancy speech tests, and binaural interaction tests (Baran & Musiek, 1991).

Dichotic Speech Tests involve the presentation of stimuli to both ears simultaneously, with the information presented to one ear being different from that presented to the other ear.

Temporal Ordering Tasks require the listener to make discriminations based on the sequence of auditory stimuli. Temporal processing is important to perception of speech and music.

Monoaural Low-Redundancy Speech Tests modify the temporal, frequency, or intensity of the acoustic signal. Children with CAPD have difficulty with closure skills when the signal is distorted.

Binaural Interaction Tests assess the ability of the CANS to process information presented to the ears. This information is presented either in a nonsimultaneous condition, sequential condition, or partial information is presented which requires the listener to integrate the partial message into a meaningful message.

THE BUFFALO MODEL

(Masters, Stecker & Katz, 1993)

AUDIOLOGICAL	SPEECH-LANGUAGE	ACADEMIC
DECODING: SLOW RESPONDERS		
Staggered Spondaic Word (SSW): Errors exceed age limits in: Right competing Left noncompeting Order effect high/low Phonemic Syntheses: Nonfused Delays < age level norms Speech in Noise: 25-40% discrepancy *Often say "huh?" or "what"*	Moderate-severe phonological disorder as a preschooler Persistent problems /l/, /r/ Difficulty comprehending morphological endings Prosodic difficulties Poor phonolgical awareness …..Problems in oral discussions, story re-telling Written discourse difficulties	Poor reader, especially phonics Poor spelling Needs repetition of directions/pausing Problems reading & writing tests
TOLERANCE FADING MEMORY: IMPUSLIVE RESPONDERS, DISTRACTED BY NOISE		
SSW: Errors exceed age limits in: Left competing Order effect high/low Ear effect low/high Smush Too quick Tongue Twister] PS: omits first sounds S/N: 40% discrepancy from quiet	Cluttering Coalescence errors Metathesis errors Poor memory for long, elaborate syntax Interrupt often *Often complain of headache and stomach aches at school or in noise; extremely fatigued at end of school day.*	Poor attention Distractible Poor reading comprehension Misses part of directions Poor handwriting
INTEGRATION: VERY LONG DELAY RESPONDERS		
SSW: Type A Pattern PS: < age limits nonfused; delays S/N: discrepancy between ears	Severe response delays, all tests Very quiet Inconstant answers to questions *Often look like severe cases of decoding and TFM*	Very poor spelling Very poor reading Very poor handwriting
ORGANIZATION: REVERSALS AND SEQUENCING PROBLEMS		
SSW: reversals PS: reversals	Begins in middle of stories Jumps around in sequence of Stories Difficulty clarifying information *Appear to have difficulty staying on task unless in one-on-one situation*	Sequencing Disorganized work Disorganized desk

Once testing is completed and information is gathered from all disciplines, a child can be placed in four different categories of CAPD described by Katz, Smith, and Kurpita (1992). These categories along with the results of audiologic, speech-language tests, and academic characteristics make up the Buffalo Model presented by Masters, Stecker, and Katz in 1993.

The Buffalo model also provides information on management strategies that are appropriate for the four profiles listed above. These management strategies focus on the child's strengths and weaknesses to improve performance across all areas. Above is a brief description of the area of focus for each category.

Decoding – improving phonemic and metaphonological skills.

Tolerance-fading memory-improving the signal-to-noise ratio and strengthening short-term memory skills.

Integration – improving phonemic and metaphonologic skills.

Organization – improving sequencing skills and organizational habits.

Other methodologies can be used to treat CAPD depending on each child's areas of weakness. A few of these methodologies include:

1. Auditory training for those children with neuro-maturational lag.
2. Metacognitive strategies which benefit listening and comprehension of spoken language.
3. Memory and attention strategies focusing on where the breakdown occurs in memory and attention and choosing appropriate strategies to remediate these deficits.
4. Frequency Modulated (FM) technology which is utilized for those children who have difficulty listening in background noise.
5. Speech and language therapy focusing on the weaknesses described in the four categories of the Buffalo Model: decoding, tolerance-fading memory, integration and organization.

There is also a list of general modifications that can be made in the classroom to facilitate a better listening environment as well as ensuring academic success. These modifications include: providing a notetaker, giving direct instructions, pre-teaching lessons, using an organizer/assignment books, preferential seating, reducing noise in the classroom, facilitating direction following, and allowing test modifications.

CAPD can have a significant impact on a child's academic success. Understanding this disorder and it's implications can assist a physician in making a more informed decision for their patient's future success. Identifying children with CAPD versus ADHD and/or both will ensure the child's needs are being identified and met.

BIBLIOGRAPHY

American Speech-Language-Hearing Association; Task Force on Central Auditory Processing Consensus Development (1996). American Journal of Audiology, 5 (2), 41-54.

Baran, J.A., & Musiek (1991). Behavioral assessment of the central auditory nervous system. In W. Rintelman (Ed.), Hearing assessment (2nd ed.) pp 549-602. Austin, TX: PRO-ED.

Bellis, T.J. (1997). Assessment and management of central auditory processing disorder in the educational setting: From science to practice. San Diego: Singular Publishing, Inc.

Katz, J., Smith, P., & Kurpita, B. (1992). Categorizing test finding in children referred for auditory processing deficits. SSW Reports, 14, 1-6.

Masters, M. G., Stecker, N.A., & Katz, J. (1993, November). CAP disorders, language difficulty, and academic success: A team approach. Presented at the American Speech-Language-Hearing Association Convention, Los Angeles.

Masters, M. G., Stecker, N.A., & Katz, J. (1998). Central auditory processing disorder: Mostly management. Needham Heights, Ma: Allyn & Bacon.

Tillery, K.L. & Keller, W. (1999, November). Comorbidity of CAPD and attention deficit hyperactivity disorder (ADHD). Presented at the American Speech-Language-Hearing Association Convention, San Francisco, CA.

Esotropia (Crossed Eyes)

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Introduction and Epidemiology

Strabismus is an ocular misalignment (Figure 1) that occurs in nearly 2 percent of American children 1 to 3 years of age and nearly 3 percent of school-age children, adolescents and young adults 13 to 24 years of age.

Figure 1

Esotropia is a type of strabismus in which one or both eyes cross or turn inward. Eso means in and tropia means to turn. It is the most common strabismus of childhood, accounting for approximately 60 percent of strabismus cases.

If a child crosses one eye more than the other, this may be a sign of amblyopia or "lazy eye". Amblyopia, commonly associated with esotropia, refers to poor vision in a drifting eye because it is not being used enough. Patching of the stronger eye may be required to correct the vision in addition to

surgery to straighten the eyes. The worse the vision, the more patching that will be required to correct the vision.

Prompt treatment of esotropia reduces both its prevalence and morbidity by increasing the likelihood of developing and/or maintaining binocular vision. Prevent Blindness America has estimated that the early identification and treatment of children with strabismus could have prevented amblyopia in approximately 5 million Americans. Certain pediatric populations are at higher risk for developing strabismus, including neurodevelopmentally impaired, premature or low birth weight infants, infants with low Apgar scores, children with craniofacial anomalies, and those with a family history of strabismus.

Types of Esotropia

There are two main categories of esotropia. The first main type known as infantile esotropia appears very early in life, usually by 6 months of age. The second main type of esotropia is acquired esotropia and appears later on, any time from age 6 months to 8 years of age, but most typically between ages 2 to 5 years old.

Infantile Esotropia

Infantile esotropia is defined as a crossing of the eyes with an onset usually by 6 months of age and an absence of signs which would suggest cranial nerve palsies, intraocular disease, or congenital aberrations of innervation. The crossing may initially seem intermittent or small, but usually soon becomes a constant crossing of the eyes which is frequently fairly large in magnitude as seen in Figure 2.

Most of these infants will cross both eyes equally, but occasionally they will tend to cross one more

Figure 2: Infantile esotropia

frequently or severely than the other. The etiology of infantile esotropia is unknown, but it runs in families and there is a genetic predisposition. The treatment for infantile esotropia is almost always surgical. This involves loosening or weakening the inner (medial rectus) eye muscles in order to straighten the eyes. This type of esotropia can almost never be corrected with glasses alone.

Acquired Esotropia

Acquired esotropia occurs in children whose eyes are straight for at least the first 6 months of life but then develop crossing after this. Children with this condition are assumed to have had normal visual development before the onset. Most cases are caused by accommodative disorders and have an onset between the ages of 2 and 5 years.

In many children with acquired esotropia the crossing is caused by a refractive error or farsightedness and can be corrected by prescribing glasses to correct the farsightedness for the child. Farsightedness (images are more blurry at near than distance) causes the eyes to cross because the child has to strain and focus to see, especially at near targets.

Glasses eliminate the need to focus and thus corrects the crossing as shown in Figure 3. This type of esotropia is also called accommodative esotropia

Occasionally bifocal glasses will be prescribed if the eyes cross worse when looking up close than when looking far away. Even if the glasses do correct the crossing, the eyes will still cross when the glasses are not being worn.

In some cases of accommodative esotropia glasses will not full correct the crossing and surgery on the eye muscles may be required. Surgery is more likely in children who have had crossing of the eyes for a long time, and in children who have developed amblyopia or “lazy eye”. Amblyopia occurs more frequently in acquired esotropia than in infantile esotropia. If it is present, patching of the stronger eye will be required in addition to glasses and/or surgery. Most children whose eyes are straightened with glasses will require glasses throughout childhood, although many of them may out grow the glasses when they reach 7 to 8 years of age. At any time during childhood if the glasses are no longer straightening the eyes, then surgery may be recommended.

There are additional causes of acquired esotropia which can be caused by neurologic, genetic and/or structural abnormalities of the eyes which can cause secondary esotropia Some of these causes include congenital cataracts, TORCH infections, optic nerve abnormalities, sixth nerve palsies, myasthenia gravis, and trauma. It’s also important to note that 25 percent of patients with retinoblastoma present initially with strabismus and not necessarily obvious leukocoria (whitish retinal reflex).

Screening

Screening for esotropia includes a brief history, and assessment of visual acuity and

Figure 3: Accommodative esotropia

testing of ocular alignment. Asymmetric or reduced vision is associated with strabismus. An ocular history of prematurity, development, vision, ocular alignment, and family history of strabismus and amblyopia should be recorded. Patient with risk factors for strabismus as previously discussed should be referred to pediatric ophthalmology for further evaluation.

Visual acuity should be tested in an age-appropriate fashion. For infants and toddlers under 3 years of age, qualitative assessments of fixation and tracking (particularly to the parent's face or interesting objects) is a valuable screen. Acuity in verbal, preliterate children age 3 to 5 years can be tested using letters, figures, "tumbling E", and in the literate child using Snellen letters. Referrals should be made to pediatric ophthalmology for an acuity of 20/50 or worse in a 3-year-old, 20/40 or worse in 5-year-olds and 20/30 or worse in older children. Any asymmetry between the eyes should also be referred.

Screening for esotropia can be accomplished by observation or by using the corneal light reflex test. By simple inspection, and examiner can identify a large to moderate esotropia and pseudoesotropia (common because of infantile broad epicanthal folds). The corneal light reflex test is administered to a child of any age by shining a penlight centrally from the examiner's face toward the child's face while the infant/child is looking at a near target. The tester evaluates the symmetry of the light reflex on the cornea; temporal displacement of the light reflex (as seen in Figures 1, 2, and 3) supports an esotropia.

Conclusion

Esotropia is the most common type of strabismus (ocular misalignment). The two most common types are infantile esotropia usually presenting the first 6 months and requiring surgery and acquired esotropia presenting after 6 months of age usually treated with glasses and/or surgery. Our goal is to promote regular visual screening to identify patients at risk for esotropia (crossed eyes), promote early detection of esotropia and amblyopia in order to restore and maintain binocular vision. Please refer any patient with any question of crossed eyes and/or decreased vision to Pediatric Ophthalmology for further evaluation and consultation.

Reference

Esotropia: Preferred Practice Pattern; The American Academy of Ophthalmology Publication, 1997.

Congenital Hypothyroidism: Screening, Diagnosis, and Treatment

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Thyroid hormone is a vital hormone for normal growth and development. Congenital hypothyroidism occurs when an infant is born without the ability to make normal amounts of thyroid hormone. If untreated, congenital hypothyroidism will result in severe mental retardation and growth failure.

Historical Perspectives

In 400 BC, sculptures of goitrous dwarfs in South America are some of the first that document the effects of untreated congenital hypothyroidism. The ancient Romans wrote about goiters in the first century AD and Paracelsus further described a picture of mental retardation and goitrous hypothyroidism in lectures and publications during the sixteenth century. It wasn't until the late 1800s that thyroid extract was reported to be an effective treatment for congenital hypothyroidism. In the 1970s it became possible for mental retardation caused by congenital hypothyroidism to be erased by the early treatment as a result of neonatal screening.

Thyroid Gland Development

During early gestation the thyroid gland begins as an epithelial proliferation and invagination of the foregut endoderm. The thyroid then migrates caudally, remaining connected to the floor of the foregut by the thyroglossal duct. Normally this duct will disappear completely, and by 12 weeks thyroid embryogenesis in almost entirely complete. This gland is able to concentrate iodine and synthesize thyroid hormone. Also by 12 weeks, the fetal pituitary gland is identifiable and contains thyroid stimulating hormone (TSH). The hypothalamic and pituitary portal vascular system begins development at about 6 weeks and is finalized at about 35 weeks gestation.

The exposure of the infant to the extrauterine environment leads to an acute release of TSH that in turn stimulates a prolonged release of thyroid hormones. The serum TSH peaks over about the 30 minutes after birth, and then rapidly decreases over the next 24 hours. The serum thyroxine (T4) levels increase to peak values at 24 to 36 hours post partum and then slowly fall over the first few weeks of life. In the preterm infant, delivery occurs before the hypothalamic-

pituitary-thyroid system is mature. These infants have a similar TSH surge and T4 rise as do term infants, however the amplitude of this response is blunted. The remainer of this article will focus on the term infant with abnormal newborn screen results, but one should remember that the incidence of congenital hypothyroidism is the same for preterm infants as it is for infants born at term.

Screening

The Texas Department of Health and Newborn Screening Program began screening for congenital hypothyroidism in February of 1980 and is the largest in the world in terms of total number of samples processed. The incidence of congenital hypothyroidism in Texas is approximately 1 in 2,500 newborns screened (the national incidence is about 1 in 4,000).

Why do we screen? The clinical diagnosis of congenital hypothyroidism occurs in fewer than 5% of newborns with the disorder, because the signs and symptoms are subtle. Without prompt intervention these children will develop growth failure, irreversible mental retardation as well as a variety of neuropsychologic deficits (known as cretinism). Early treatment leads to normalization of the growth potential and prevention of the significant mental retardation.

How does the screening process work? The first step in the screening process is the collection of blood from a heel puncture onto filter paper. This should be done a minimum of 24 hours after birth, in order to avoid the post-natal TSH surge (a second screen is required at one to two weeks of age). Most of the laboratories in the United States measure T4 levels, however in several states and in most of Europe TSH is the initial screening test. The sample is sent to the State Laboratory in Austin where the T4 level will be measured. Samples with the lowest 10% of T4 will be repeated along with a TSH. The lowest 0.5% of those samples will be reported as abnormal. Abnormal tests are not diagnostic and must be confirmed with venipuncture blood samples.

Interpretation of Results

There are a variety of conditions which will lead to low T4 levels in newborns. See Table 1. These diagnoses can normally be distinguished by a thorough history, physical and appropriate laboratory studies. See Table 2.

Primary hypothyroidism is most frequently caused by abnormalities in the formation, migration or growth of the thyroid gland (termed thyroid dysgenesis). This accounts for 75-85% of the infants with congenital hypothyroidism. These defects occur in a sporadic fashion and the exact cause is unknown. Another cause of primary hypothyroidism is an organification defect; that is an abnormality in one of the enzymatic reaction steps involved in the production and release of thyroid hormone (termed thyroid dyshormonogenesis). Thyroid dyshormonogenesis accounts for about 10-15% of infants with congenital hypothyroidism. These defects are inherited in an autosomal recessive fashion.

Secondary or central hypothyroidism is due to hypothalamic or pituitary dysfunction. The thyroid gland is properly formed and in its normal location, but TSH is either not produced or is not released from the pituitary gland. This leads to lack of thyroid hormone production. Fewer than 5% of infants with congenital hypothyroidism will be in this category.

Clinical Presentation

Most hypothyroid infants have no symptoms before discharge from the nursery. Clinical hypothyroidism generally does not appear until 2 or 3 months of age. On occasion an infant with thyroid dysgenesis will have a normal newborn screen and will present later in childhood with signs and symptoms of hypothyroidism. Untreated infants with hypothyroidism may have hypothermia, poor feeding, bradycardia, jaundice (unconjugated hyperbilirubinemia), enlarged posterior fontanel, or an umbilical hernia. An infant can have congenital hypothyroidism despite having a normal newborn screen, so if you suspect hypothyroidism based on an infants clinical presentation draw thyroid function studies.

Diagnosis and Treatment

The evaluation of an infant with an abnormal newborn screen

TABLE 1.

Primary Hypothyroidism
Secondary Hypothyroidism
Low Thyroid Binding Globulin Levels
Maternal Medications (iodides, lithium PTU)
Prematurity
Significant Stress or Illness
Idiopathic Transient Hypothyroidism
Maternal Thyroiditis

*Adapted from Texas Department of Health-Hypothyroidism website

TABLE 2.

Screen Results	Possible Cause
↓T4 / ↑ TSH	Primary Hypothyroidism
	Maternal Antibodies
	Maternal Medications (PTU, Iodine)
↓T4 / NL TSH	Secondary Hypothyroidism
	Prematurity
	Thyroid Binding Globulin Deficiency
NL T4 / ↑ TSH	Secondary Hypothyroidism
	TSH Surge

should include a careful history, physical examination, and serum confirmation of the thyroid hormone levels. The recommended laboratory studies to obtain are TSH and free T4. If your lab does not do free T4, total T4 and serum TBG levels can be obtained instead. If the TSH is markedly elevated, prompt treatment should be instituted while waiting for confirmatory lab results. DO NOT WAIT FOR THE CONFIRMATORY RESULTS TO BEGIN THYROID HORMONE REPLACEMENT. The goal of treatment is to achieve normal serum T4 and TSH levels as quickly as possible, maintain them in the normal range thereby allowing the infant to have normal growth, development, and intellectual function. The recommended treatment is oral L-thyroxine therapy at 10 – 15 mg/kg/day (term infants are usually between 25 – 50 mg/day). Repeat thyroid function studies should be performed 2 – 4 weeks after starting therapy and at a minimum of every 3 months during the first year of life.

It is important to treat infants with borderline hypothyroidism. Thyroid hormone is critical for normal brain development, especially during the first 2 years of life. It is recommended that an infant who has borderline hypothyroidism remain on thyroxine replacement until the age of 3 years, when a trial off of thyroid hormone replacement may be considered.

It is important for the health care provider to instruct the parents on how to properly administer the medication. L-thyroxine comes in pill form. The pill should be crushed in a small amount of water or formula. Soy formula, high iron containing formulas, iron supplementation, and citrus juices interfere with the absorption of thyroxine. Babies on soy formulas may need a higher dose of thyroxine than an infant not on soy formula. If the baby is on iron, thyroxine should not be given at the same time as the iron supplementation. The acid from citrus juices will dissolve and inactivate the thyroxine, so care should be taken not to administer the thyroxine in conjunction with orange juice or other acidic juices.

Summary

Congenital hypothyroidism is a common disorder seen by health care providers who take care of infants. The health care provider must be able to interpret the results of the newborn screen, obtain confirmatory testing of thyroid function, and begin appropriate therapy in infants suspected of having congenital hypothyroidism. Quick and accurate attention to the infant with an abnormal screen will allow that infant to have the best possible outcome.

Bibliography

1. Foley, Thomas P. Jr. Congenital Hypothyroidism. Werner and Ingbar's The Thyroid. 7th Edition, edited by Braverman LE and Utiger RD. 1996. Pages 988 – 993.
2. Texas Department of Health. Congenital Hypothyroidism – Guidelines for Health Care Professionals. www.tdh.state.tx.us/newborn/c_thyro.htm.
3. Pass, KA, Lane PA, Fernhoff, PM, et al. US Newborn Screening System Guidelines II: Follow-up of Children, Diagnosis, Management, and Evaluation. Statement of the Council of Regional Networks for Genetic Services (CORN). 2000 J Pediatrics 137 (4).
4. Hopwood N. Managing the Patient with Congenital Hypothyroidism. Clinical Update – Pediatric Endocrinology. Summer 2000, pages 6 – 7.

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Our goal as part of the pediatric team is to provide quality imaging while maintaining a safe environment for the patient. Any questions or concerns can be addressed by any of the pediatric radiologists. Important numbers are listed below.

Scheduling: 292-6085 for routine exams.
US: 292- 7164 (Pediatric tech: Susan)
Fluoroscopy: 292-3738 (Pediatric tech: Sue)
Reading room: 292-5207

Dr Richardson: Pager – 292-6110 (2720)
Dr Keesling: Pager – 292-6110 (1109)
Dr Taylor: Pager – 713-2638

Welcome to Research Corner!

This is a quarterly addition to the SAMPC Newsletter intended to highlight research activities in the military pediatric community and promote a research friendly environment. If you have an approved protocol related to the health of children or families we would like to post it here under **Ongoing Research**. This will also be a forum to find research ideas, collaborators, and mentors (**The IDEA Board)**. We will be posting reminders for upcoming research meetings and submission deadlines. If you have other suggestions or would like to post a protocol, idea or meeting announcement in this space, please contact MAJ Cydney Fenton, SAMPC Research Coordinator at 292-7276 or Cydney.Fenton@59MDW.WHMC.AF.MIL.

Ongoing Research

*Asthma informatics – Validation of HEDIS 3.0 metric – COL Inscore (916-0707)
*Ask-a-Doc Regional referral system validation – COL Inscore (916-0707)
*Telemedicine Grant – three different studies – COL Inscore (916-0707)
*POG chemotherapy protocols – LTC Barker (292-6689)
*Bovine polymerized HGB use in ECMO in a sick pig model – MAJ Wilson (292-6625)
*Incidence of Major Congenital Malformations – DACH v WHMC v the World – MAJ Wilson (292-6625)
*Validation of Neonatal PFT machines/Ventilators using a test lung – MAJ Wilson (292- 6625)
*Neonatal ECMO Transport in the 1990s – Outcomes/Complications v Inpatient – the WHMC Experience – MAJ Wilson (292-6625)
*Case Report – CDH in patient with 7q- - MAJ Wilson (292-6625)
*Comparisons of newborn health outcomes comparing public, private, and military hospitals in TX – COL Heiman (292-7446)
*Prevalence of Genetic Disease in Pediatric Inpatients at DACH and WHMC – LTC McLean (292-7750) and MAJ Croley
*Evaluating various techniques of capillary refill in newborns – COL Heiman (292-7446)
*Use of growth hormone for nutritionally deplete children with CF – COL Inscore, MAJ Fenton, and CPT Schobitz

IDEA Board

If you are interested in working on any of the following please call the POC.
*Depo-Provera – side effect profile – MAJ Ahrendt (916-2258)
*Hirsutism Scale – effect of ethnicity – LTC Dillon (916-3530)
*Adenosine induced atrial fibrillation – case report – MAJ (P) Malpass (292-2068)
*Utility of pre-discharge pulse oximetry in the newborn nursery: Can we detect subtle, serious CHD before early discharge? – MAJ (P) Malpass (292-2068)
*Cardiac ausculatory skills in pediatric housestaff: can we teach residents to become better listeners? MAJ (P) Malpass (292-2068)
*Telemedicine research – COL Heiman (292-7446)
*Research and equipment for neonatal transport – COL Heiman (292-7446)
*Use of the internet in managing diabetics - does it improve control and follow-up? – MAJ Fenton (292-7276)

Useful Numbers

BAMC Department of Clinical Investigation – 916 – 3511
WHMC Clinical Investigation Department – 292 - 7141

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